

# Dandy Walker Complex

*Patient Information Series – What you should know, what you should ask.*

## **What is dandy walker complex?**

Dandy walker complex is a group of overlapping conditions involving the back (posterior compartment) of the brain and the cerebellum, the part of the brain responsible for coordinated movement. These conditions share in common certain features on ultrasound, that give the impression that one of the fluid-filled spaces in the brain (the 4th ventricle) is in communication with the most posterior space in the brain, the cisterna magna. It includes many conditions such as Dandy walker malformation, Vermian hypoplasia, agenesis, Blake's pouch cyst, Joubert's syndrome etc. These disorders are grouped together as they are often difficult to differentiate and they tend to overlap clinically.

## **How do these structural differences in the brain develop?**

This constellation of conditions develops late in the first trimester, when the posterior brain structures are forming, for reasons not quite clear. Some of the conditions, such as Blake's pouch cyst likely represents a delay in normal development and usually resolves later in the pregnancy. More severe conditions in this spectrum, involving the cerebellum, such as Dandy Walker malformation, are linked to abnormal development of brain structures and have more severe consequences for the baby.

## **How are these conditions diagnosed?**

Ultrasound is the mainstay of diagnosis. At each routine survey of fetal anatomy performed in the mid-trimester, the fetal brain is examined. If there is a suspicion that structures in the posterior compartment of the brain appear unusual and Dandy Walker Complex is suspected, you may be referred for a more detailed fetal brain scan, called fetal neurosonography. In this examination, in addition to the standard views of the baby taken through your abdomen, a transvaginal scan may be advised if the baby is in a head-down position to give more detailed views of the baby's brain anatomy. Finally, a fetal MRI may be recommended to give further views of the back of the brain and surrounding structures. The combination of these imaging investigations can differentiate these conditions and help arrive at a definitive diagnosis.

## **What are the genetic or chromosomal implications of diagnosis of Dandy Walker Complex?**

The Dandy-Walker complex is frequently associated with differences in the chromosomes and genetic testing is advisable. In addition, many syndromes and other brain malformations can be found in association with these disorders.

## **Should I have more tests done?**

As indicated above, a detailed Neurosonogram is advised and an MRI may be suggested. A detailed ultrasound evaluation is needed to rule out other structural differences beyond the brain. Serial ultrasounds will be suggested to assess evolution of the findings. Genetic counseling and invasive fetal genetic testing, in the form of amniocentesis maybe offered to rule out chromosomal or other genetic disorders.

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## **What are the things to watch out for during the pregnancy?**

Care during ongoing pregnancy is not likely to be significantly affected when these conditions are diagnosed. These conditions generally do not warrant early delivery. Vaginal delivery is not contraindicated except in cases where there has been enlargement of the fetal skull due to excess fluid in the brain cavities. Your baby should be delivered in a centre with access to detailed imaging services and input from paediatric specialists, particularly a paediatric neurologist and/or neurosurgeon to plan for investigation and care after birth.

## **What does this mean for my baby after birth?**

Prognosis in these conditions is highly variable and largely dependent on specific diagnosis. Generally if isolated and in the presence of normal chromosomes, the prognosis in Blake's Pouch cyst is excellent. More severe involvement of the cerebellum, such as is found in Dandy Walker Malformation can be associated with neurodevelopmental impairment, particularly if associated with chromosomal or other abnormalities. Joubert's Syndrome is often lethal or associated with severe intellectual impairment.

## **What other questions should I ask?**

- Is my baby's diagnosis Blake's pouch cyst, Dandy walker malformation, or Joubert's syndrome?
- Are other structural differences present?
- What genetic testing is available?
- How often will I have ultrasound examinations done?
- Where should I deliver?
- Do I need a Caesarean Section?
- Can I meet the doctors caring for my baby after birth?

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